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Joint Hypermobility in Children Referred to Rheumatology Clinics

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Abstract: The term benign hypermobility syndrome is applied to those children with musculoskeletal pain associated with generalized hypermobility of the joints without any associated congenital syndrome or abnormality of connective tissue, such as Marfan or Ehlers-Danloss syndrome. The aim of this study, was to determine the prevalence of joint hypermobility among school students and to define the characteristics of patients with joint hypermobility. This study was conducted between January 1994 and July 2004 among school students in Tehran. The clinical features were often associated with intermittent nocturnal pains and are characterized by the occurrence of musculoskeletal symptoms in the absence of demonstrable systemic rheumatologic disease. The degree of joint was scored by modified criteria of Carter and Wilkinson. Twenty hundred fifty two students (132 females and 120 males) with a mean age of 8.7 years (range 6-16) were examined. Joint hypermobility was found in 52 children (20.5%), but Joint hypermobility syndrome was observed in 30 (11.8%) of the students. There were 12 male (40%) and 18 female (60%) hypermobile subjects. Although hypermobility does not seem to be very problematic in young people, as in our focus group, we believe that it is important for physicians to recognize this problem to ensure correct diagnosis and treatment, since it may lead to mimic rheumatic diseases in the future.

Key words: Beighton scale, Carter and Wilkinson, joint hypermobility, joint laxity, nocturnal pain, children

INTRODUCTION

The hypermobility syndrome in children has been described by Kirk *et al.* (1967). The diagnosis of hypermobility is made clinically, without need for costly or invasive testing (Grahame, 1994). Joint hypermobility is defined as abnormally increased mobility of small and larger joints beyond the limits of their physiological movement (Grahame, 1990). The term benign hypermobility syndrome is applied to those children with musculoskeletal pain associated with hypermobility of the small and large joints. A hypermobile joint is one whose range of movement exceeds the norm for that individual, taking age, sex and ethnic background into consideration.

The hypermobility syndrome musculoskeletal pain of non-inflammatory origin is a common cause of morbidity in childhood. The hypermobility syndrome is often associated with intermittent nocturnal pains that may occur after certain activities and these disorders are characterized by the occurrence of musculoskeletal symptoms in hypermobile subjects in the absence of demonstrable systemic rheumatologic disease (Grahame, 1978).

The criteria for diagnosis of this disorder mostly used nine-point Beighton scale or the modified criteria of Carter and Wilkinson (Grahame, 2000). The frequency of hypermobility is estimated from 8-20% depend on age, criteria for assessing hypermobility and ethnicity (Biro *et al.*, 1983; Gedalia *et al.*, 1985; Bridges *et al.*, 1992). Caucasian and West Africans have a much higher prevalence of hypermobility (Adib *et al.*, 2005; Grahame, 1990; Bightonl *et al.*, 1973; Seçkin *et al.*, 2005). Girls are affected more than boys and this entity is common in young women, with an estimated prevalence of 5% in the healthy adult population.

The purpose of this study, was to determine the prevalence of joint hypermobility among school students and to define the characteristics of patients with joint hypermobility.

MATERIALS AND METHODS

This study was conducted among school students between January 1994 and July 2004 in the Tehran. All subjects had been referred to pediatric rheumatologic clinic for muscle-skeletal pain. In all students, we

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performed complete history and physical examination. In order to designate marfanoid feature and Ehlers-Danloss syndrome, body weight, height, span/height, upper/ lower segment ratios and skin examination were recorded and Marfan or Ehlers-Danloss syndrome were excluded in all subjects by physical examination. The criteria for diagnosis of this disorder mostly used nine-point Beighton scale or the modified criteria of Carter and Wilkinson (Table 1). The degree of joint was scored on a scale between 0 and 9 using the method described by modified criteria of Carter and Wilkinson. A score of 4 or more indicates benign joint hypermobility syndrome or generalized hypermobility of the joints (Grahame, 2000). For defining the characteristics of patients, the following features were also examined: arthralgia, myalgia, low back pain, sciatica, spinal deformities. Statistical analyses were performed using the Statistical Package for Social Science program (Version 11).

RESULTS

In this study, 252 school students were examined, including 120 males (47.6%) and 132 females (52.4%). The mean age of our subjects was 8.7 years (range 6-16). Joint hypermobility was found in 52 children (20.5%), but 30 (11.8%) had the criteria of benign joint hypermobility syndrome by the Bighton or Carter and Wilkinson criteria (score more than 4). These cases were included 12 male (40%) and 18 female (60%) subjects (female/male ratio:1.5/1). The mean age of these groups was 9.1 years (range 6-16). Mean age of this group in sex distribution was 8.7±1.1 years for males and mean age for females was 9±1.1 years. Table 2 shows frequency of the hypermobile students in different ages. The age, weight and height of children with hypermobility were lower than normal children. These characteristics of the normal and hypermobile students are summarized in Table 3 and 4 shows clinical manifestations in hypermobile students.

Table 1: Bighton hypermobility score

	Clinical symptoms*	Right	Left
1	Passively dorsiflex the fifth metacarpophalangeal joint to >90	1	1
2	Oppose the thumb to the volvar aspect of the ipsilateral forarm (Fig. 1)	1	1
3	Hyperextend the elbow to >10	1	1
4	Hyperextend the Knee to >10	1	1
5	Place hands flat on the floor without bending the Knee	1	
	Total	9	

*One point can be had for each side (for maneuvers1-4), so there score will have a maximum of nine parts if all are positive]

Table 2: Frequency of the hypermobile students according to the Bighton scoring system

Age	Frequency
6/y	5 (10%)
7/y	6 (12%)
8/y	7 (13%)
9/y	7 (13%)
10/y	7 (13%)
11/y	5 (10%)
12/y	5 (10%)
13/y	3 (5%)
14/y	3 (5%)
15/y	2 (4%)
16/y	2 (4%)
Total	52 (100%)

Table 3: Characteristics of the normal and hypermobile students

	Normal	Hypermobile
Characteristics	Mean (SD)N=200	Mean (SD)N=52
Age (year)	10 (2)	9 (2)
Body weight in (kg)	40 (5)	30 (4)
Body height (cm)	150 (6)	130 (5)
Span/height ratio	1 (-0.04)	1 (-0.03)

Table 4: Clinical manifestation of hypermobile students

Symptom	Frequency
Arthralgia	20 (38%)
Myalgia	18 (34%)
Low back pain	16 (30%)
Joint sprain	14 (26%)
Joint dislocation	12 (23%)
Joint Effusion	8 (15%)
Sciatica	6 (11%)

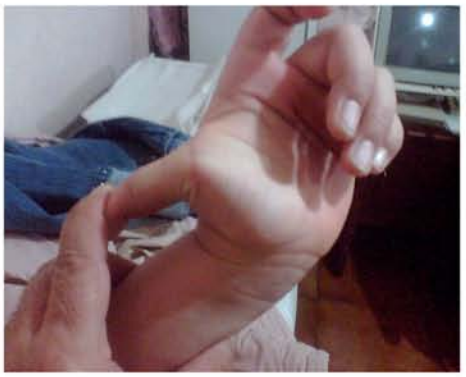

Fig. 1: Oppose the thumb to the volvar aspect of the ipsilateral forearm

DISCUSSION

Although not studied in a formal fashion, it is our impression that a few children have slightly joint hypermobility. Studies on populations show that joint hypermobility is observed more often in children, aged 6-16 years are mostly affected, as the prevalence decreases with age (Al-Rawl *et al*., 1985) and with increasing age, hypermobility syndrome decreased (Seçkin *et al*., 2005). Our subjects were age 5-16 years old and frequency of hypermobility syndrome increased up to 8 years old and then decreased.

The criteria for hypermobility have evolved over the years and currently most authors use either the nine point

Beighton scale or the modified criteria of Cater and Wilkinson (2000). Generalized joint hypermobility is a feature of inherited disorders of connective tissue such as, Marfan's syndrome and Ehler-Danlos syndrome and marfanoid features has been described in children with the hypermobility syndrome (Grahame, 1999, 2000; Moore *et al*., 2006; Yen *et al*., 2006).

Joint hypermobility was commonly observed on pediatric patients referred to pediatric rheumatologic clinics and in normal school children (Bridges *et al*., 1992; Adib *et al*., 2005). Our study showed 20% children who were referred to our clinic, had hypermobility joint and 11.8% benign joint hypermobility syndrome. The prevalence of hypermobility in the overall population varies according to sex, age and race. In Arabic populations joint hypermobility was found in 25.4% of males and 38.5% of females (Al-Rawl *et al*., 1985). Biro *et al.* (1983) found hypermobility in children with female: male ratio of 11:4 in 5.7% of the referral patients. Bridges *et al.* (1992) found hypermobility in 15% of adult patients who was referred to a rheumatologic clinic and reported that joint hypermobility may mimic rheumatic diseases. Girls generally show a greater joint range than boys (Adib *et al*., 2005; Al-Rawl *et al*., 1985; Kirk *et al*., 1967; Seçkin *et al*., 2005; Lewkonia and Ansell, 1983; Bridges *et al*., 1992; Gedalia *et al*., 1985). Our findings on the higher occurrence of hypermobility in female subjects support the earlier studies (female to male ratio 1.5:1).

Patients with joint hypermobility usually present with arthralgia, back pain, traumatic or overused soft tissue lesions, recurrent joint dislocation or subluxation, low grade inflammatory arthritis abnormal gaite (Adib *et al*., 2005, Bridges *et al*., 1992; Murray and Woo, 2001). All of children with hypermobility will have musculoskeletal disorders. The mechanism of these disorders is not understand well, but the most sign of joint laxity and associated clinical features, such as (easy bruising, slow tissue healing and skin elasticity) may indicate that inherent disorders (Remvig *et al*., 2007; Grahame, 1994). Furthermore, it can be a different and independent entity of inflammatory conditions of connective tissue or other congenital disorders (Gedalia *et al*., 1985). Some evidence shows that arthralgia, is a major component of alleged hypermobility-related problems. It is suggested, there is a correlation between osteoarthritis and hypermobility but it is not proven (Remvig *et al*., 2007). There are no randomized controlled studies regarding effects of existing treatments. All subjects in this study had a musculoskeletal complaint but arthralgia, myalgia and low back pain were the main symptom in hypermobile children. Joint effusion was seen in 15% our subjects.

A very few children will require a change of activity not be interpreted by the family or child as a recommendation to stop participating in physical activities. A benign bleeding tendency has been reported in children with hypermobile syndrome. Although hypermobility may enable a child to be good gymnast, or dancers, injuries may be more frequent in hypermobile athletes as well as in individuals undergoing strenuous training, including military recruits. According to Al-Rawl *et al*. (1986) study the rate of joint injury, ligamentous sprains and some other musculoskeletal disorders were significantly increased more frequently in children with scored 7 or more. Laxity of the joints has been suggested as a predisposing factor for joint or soft tissue injury (Grahame, 1994, 2000; Gedalia *et al*., 1985). Because synovitis of traumatic origin is often misdiagnosed as chronic inflammatory arthritis, patients and doctors may undergo unnecessary intensive diagnostic procedures (Grahame, 1994; Murray and Woo, 2001; Misra *et al*., 1996).

CONCLUSION

Although joint hypermobility does not seem to be very problematic in young children, as in our focus group, we believe it is important for physicians to recognize this problem to ensure correct diagnosis. The first step is for physicians to recognize this entity so that they and their patients will know the pain and other symptoms are neither imaginary nor neurotic in origin. Explaining joint protection methods and advising on proper occupational and severe sport activities are necessary and usually sufficient.

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